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Anesthetic management of a 13 year old child with kyphoscoliosis and complete cleft of soft palate posted for surgical repair of soft palate

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ABSTRACT

Due to the unique anatomical and physiological variations of the airway of a child with cleft lip and palate, anesthesiologists may find it difficult to manage it properly. As a result, cleft lip and palate surgery in children will necessitate careful attention, detailed understanding, and practical application in anaesthetic preparation. While the majority of individuals have isolated CLP, a considerable percentage of them also have related abnormalities. Other issues with the formation of the branchial arch are the most noticeable (e.g. ear or upper airway defects). Anomalies of the heart, kidneys, and skeleton are among them. Associated anomalies could be part of a defined pattern, indicating which flaws to expect; the number of such disorders, including CLP, is growing by the day, ranging from the obvious, such as Trisomy 21, to the more subtle, such as Velocardiofacial Syndrome. Here we report anaesthetic management of a 13 old girl with congenital complete cleft of soft palate with kyphoscoliosis posted for surgical repair of soft palate under general anesthesia.

Keywords: Cleft palate, Kyphoscoliosis, anaesthetic management.

1. INTRODUCTION

One of the most common craniofacial malformations is Cleft lip and palate (CLP), with an incidence of 1:750 for cleft lip with or without cleft palate. There is an incidence of 1:2500 for cleft palate only (Abdelsalam & Joseph, 2021). In addition, CLP account for approximately 50% of all cases, while isolated cleft lip and solitary cleft palate each account for 25% of cases (Goswami et al., 2016). The incidence is higher in Asian countries, 1 in 500-600 live birth (Maharjan, 2004). In 2008, World Health Organization included CLP in their Global Burden Disease, as these birth defects lead to significant infant mortality and childhood morbidity (Karki et al., 2017). Infants with a CLP struggle with feeding in order to maintain adequate nutritional intake (Sai

Sankar et al., 2012). More than 150 syndromes have been associated with clefts which may further complicate the anesthetic management, but fortunately all are very rare (Stanier & Moore, 2004). Out of these, Pierre Robin Sequence (micrognathia, glossoptosis, cleft palate), Treacher Collin's syndrome (hypoplasia of maxilla, zygoma and mandible), Goldenhar syndrome (Hemifacial and mandibular hypoplasia, Abnormalities of the cervical spine) are of anesthetic significance. Here we report anaesthetic management of a 13 old girl with congenital complete cleft of soft palate with kyphoscoliosis posted for surgical repair of soft palate under general anesthesia.

Congenital scoliosis is a condition in which the spine develops abnormally from birth resulting in combination of missing portion, partial formation, or lack of separation of the vertebras. Anomaly in vertebral development in the embryo causes it. (Hedequist & Emans, 2004). Congenital scoliosis affects about one out of every 1,000 births (Sparrow et al., 2012). Unlike congenital scoliosis, congenital kyphosis and kyphoscoliosis are far less prevalent. Because compression of the spinal cord and paraplegia can occur, they are potentially more dangerous. The history and physical examination of the patient are the first steps in the evaluation process, followed by the use of relevant imaging modalities (Hedequist & Emans, 2007).

Providing anesthesia for cleft surgeries itself is a difficult endeavor in and of itself. Most of the anesthetic morbidity related to these procedures relates to the airway management, either difficult intubations or postoperative airway obstruction (Jones, 1971). The patient's quality of life can be improved by early detection and care of concurrent abnormalities (Hensinger, 2009). Here we report anaesthetic management of a 13 old girl with congenital complete cleft of soft palate with kyphoscoliosis posted for surgical repair of soft palate under general anesthesia.

2. CASE REPORT

A 13 year old girl with congenital kyphoscoliosis presented with congenital complete cleft of soft palate. She was delivered by normal vaginal route at 38 weeks of gestation with weight of 2500g. She had an Apgar score of 10. History of neonatal ICU stays of 1 week in view of congenital abnormalities and jaundice. No history of any other comorbidity noted. Her immunization history is complete up to the age. On general examination, the patient was thin built with a weight of 27kg and height of 132 cm. Her pulse rate was 88/min, her blood pressure was 110/74mmHg, her respiratory rate was 18/min and she was maintaining saturation of 98-100% on room air. She was conscious, cooperative and oriented to time, place and person. No pallor, edema, icterus, cyanosis, clubbing or enlarged lymph node. She was afebrile.

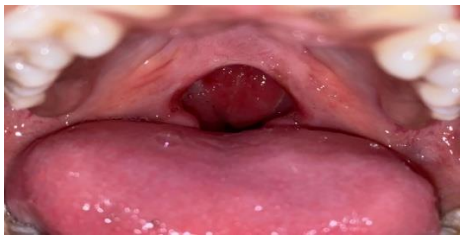

Figure 1 Cleft of soft palate

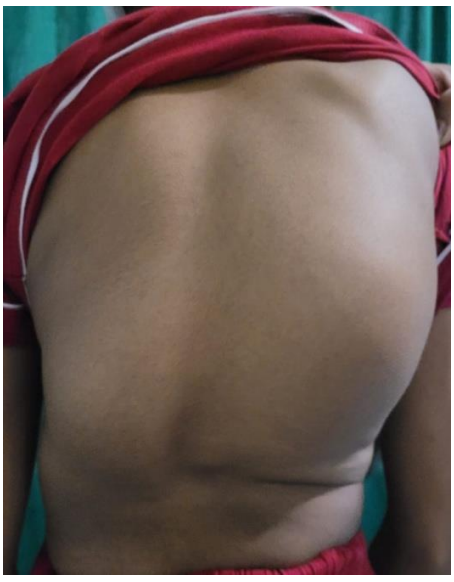

Figure 2 Kyphoscoliosis

On airway examination, she had 3 finger mouth opening with Mallampati classification of II. No loose/bucking/protruding teeth. Complete cleft palate was noticed (figure 1). Movement of neck was showing full range. TMJ movement was also normal. Spine examination showed thoraco-lumbar Kyphoscoliosis (figure 2).

On examination of respiratory system, she had normal vesicular breath sounds with equal bilateral air entry and no added sounds. No tracheal deviation was seen. There is no recent history of upper or lower respiratory tract infections. Her chest x-ray shows normal lung parenchyma (figure 3). Her Pulmnonary Function Test (PFT) showed restrictive lung pattern (FEV1/FVC >95% and FVC <44%), which can be because of kyphoscoliosis and/or due to cleft in soft palate she was unable to do proper forceful expiration (figure 4).

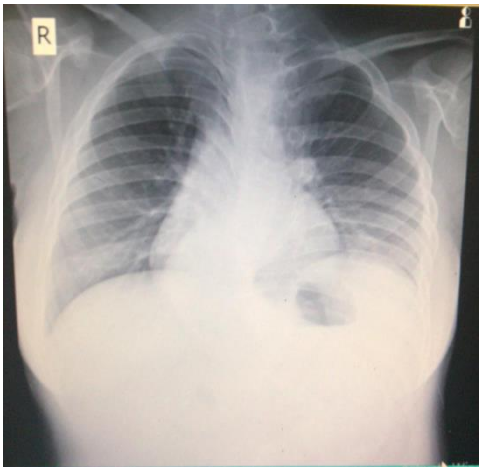

Figure 3 Chest X-ray PA view

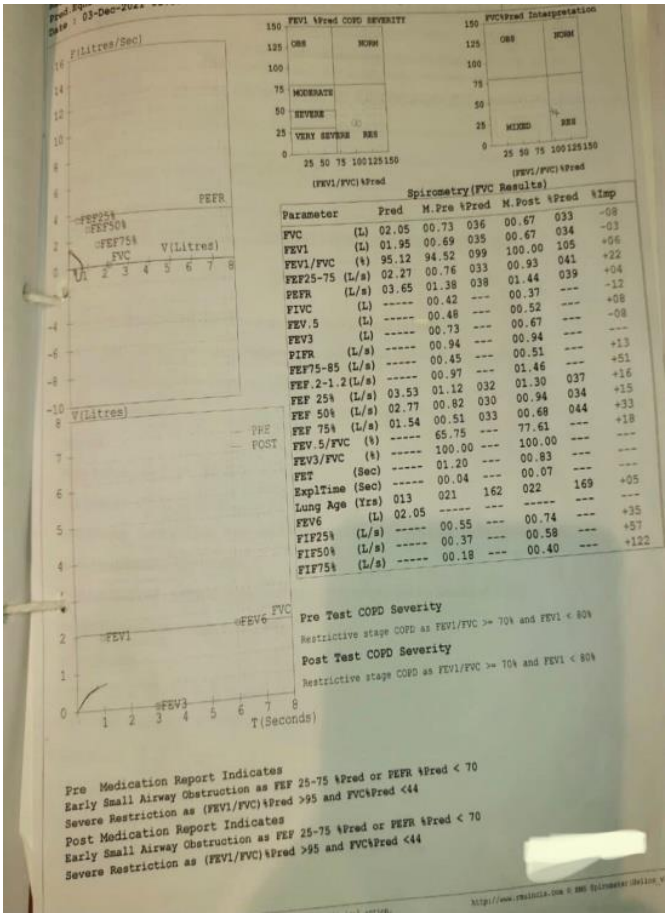

Figure 4 Pulmonary Function Test

On cardiovascular systemic examination, no visible pulsations were seen. JVP was normal. On auscultation S1 S2 heard, no murmurs heard. 12 lead ECG was taken and found to be normal and 2D echo revealed no abnormalities and showed Ejection fraction of 60%. Her CNS examination showed full GCS with no neurological abnormalities. Thoraco-lumbar spine deformity was noted and X-ray was taken (figure 5). She had no sensory or motor impairments.

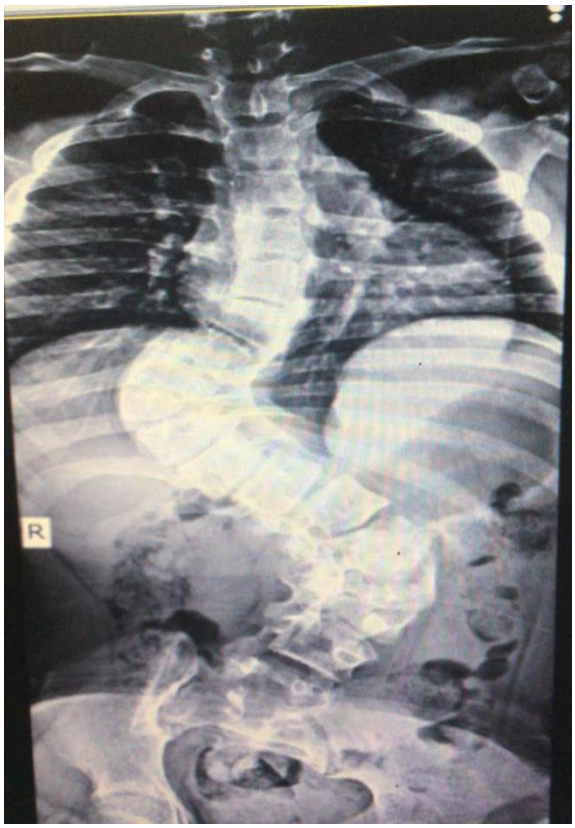

Figure 5 Xray of thoraco-lumbar spine

GIT examination was within normal limit. Menarche was attained at her age of 12 years, following which she has regular periods of 28 days with 4-5 days of bleeding associated with dysmenorrhoea. Her USG abdomen and pelvis was normal with no abnormal finding. No history of trauma in the past. No history of any drug /food/particulate allergy. No previous histories of any surgeries were noted. No history of chronic diseases or congenital anomalies in the family. Her routine blood investigations like complete blood count, kidney, liver, thyroid function tests were normal. Her liver enzymes and coagulation profile were also normal. After discussing the plan of management with the smile train surgeon and obtaining paediatric fitness for the surgery and taking opinion from Senior Cardiologist, Neurologist and Pulmonologist she was posted for an elective surgery to repair cleft of soft palate.

Anaesthetic management

Before shifting the patient to the operation theatre, NBM was confirmed, URTI/LRTI was ruled out and written informed consent was taken. After shifting to the operation theatre, patient was placed in supine position and soft roll made of cotton pads was placed in the back in such a way that her hump in the back due to kyphosis was placed in the hollow space of the roll and it helped to give a proper supine position for intubation (figure 6). After that a 20G IV cannula was secured on the left land. Standard monitors (Non-Invasive Blood Pressure, Electrocardiogram, Pulse oximetry) were attached and vitals were confirmed to be within normal limits. Inj. Amoxicillin-clavulanic acid 25mg/kg IV was given after test dose as antibiotics. IV fluids were given according to Holliday-Segar formula. Inj. Glycopyrrolate 0.004mg/kg was given 5 mins prior to induction. Oro tracheal intubation was planned with cuffed southpole tube of 6.0 or 6.5 mm. Alternative airway devices were prepared for a possible difficult airway, such as appropriately sized airways and endotracheal tubes [Ring Adair and Elwyn (RAE) tubes], Miller and Macintosh laryngoscope blades and laryngeal masks.

Figure 6 Positioning for induction

Induction of anaesthesia was done with Inj. Midaz 0.05 mg/kg IV, Inj. Fentanyl 1 mcg/kg IV, Inj. Propofol 2mg/kg IV and Sevoflurane inhalation with a MAC of 2-2.5. After confirming proper bag-mask ventilation, Inj. Vecuronium 0.1mg/kg IV was given as muscle relaxant. After preoxygenation with 100% oxygen for 4 minutes direct laryngoscopy done with Macintosh laryngoscope blade size 2, epiglottis visualized (Cormack-Lehane grade I) and oro tracheal intubation done easily with South Pole 6.5 mm tube (cuffed) that comes out over the lower lip and is secured in the centre, providing for good surgical access. Controlled ventilation was provided with fresh gas flow of 4L/min (O2 and N2O at 2L/min each). Inj. Hydrocortisone 2mg/kg was given after intubation. Throat packing was done. For the neck extension to be proper, a head ring was kept beneath the occiput also made a roll of cotton which was kept underneath the shoulders. The eyes were protected (figure 7).

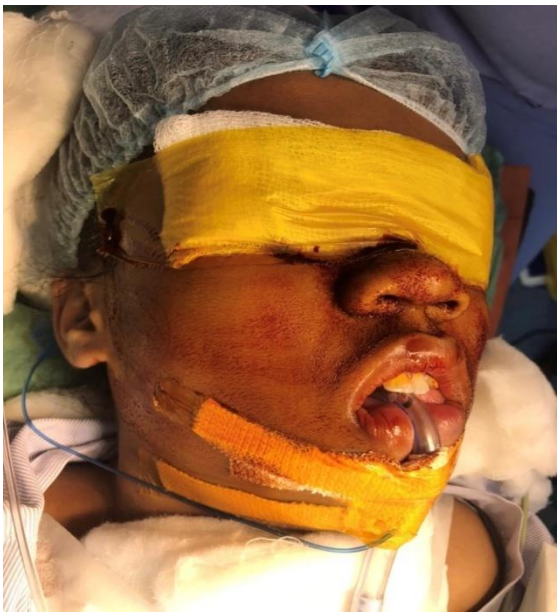

Figure 7 Post intubation

Patient was put on the ventilator on controlled mode of ventilation at a tidal volume of 8-10 ml/kg and respiratory rate of 16-18 /min and anaesthesia was maintained with volatile anaesthetic agent Sevoflurane at a MAC values of 2-2.5 and muscle relaxant topups with Inj. Vecuronium 0.5mg IV. Inj Fentanyl 0.5 -1 mcg/kg IV was given to provide analgesia intraoperatively in addition to local anaesthetic infiltration at the surgical site. Bilateral greater palatine nerve blocks were given. Strict monitoring of vitals and airway pressures were done, since the probability of tube kinking during procedure is very high. Warming blankets were provided to prevent hypothermia.

After the surgical procedure was completed thoroughly inspected the oropharynx, removed the throat pack as well as blood clots and haemostasis confirmed. Extubation was planned after switching off the inhalational anaesthetics and N2O, 100%O2 was

given. Proper elimination of volatile anaesthetic agents confirmed after spontaneous respiratory efforts, the patient was reversed with Inj. Neostigmine 0.05mg/kg IV and extubated after being fully awake. Oxygen supplementation was given. Signals of airway obstruction were not seen and chest auscultation showed bilateral equal air entry with no added sounds. The patient was given left lateral position, Inj. Paracetamol 15mg/kg IV was given for post operative analgesia. Patient was shifted to post operative room after confirming 100% SPO2 in room air and no bleeding was seen.

3. DISCUSSION

Anaesthetic management of paediatric patients itself is challenging. It will require keen and proper evaluation if associated with congenital anomalies and comorbities. A comprehensive team of experts must manage children with cleft palates. A high index of suspicion for conditions in relation to CLP should be maintained. Children with CLP should be evaluated for other anaomalies. VACTERL anomalies (Vertebral, Anal atresia, Cardiac defects, Tracheo-Esophageal, Renal & Limb anomalies) should be ruled out (Stanier & Moore, 2004).

Children with CLP are more prone for respiratory tract infection (URTI/LRTI), hence before planning for surgery any infections should be ruled out. URTIs are very frequent, and they come with a higher risk of airway problems and poor wound healing (Takemura et al., 2002). Despite the fact that many of these children have a constant nasal discharge without an infection, routine or targeted antibiotic therapy for low-grade nasal infections reduces the risk of postoperative pyrexial illness. Anemia can be caused by a combination of dietary and physiological factors. Preoperative visits should enable time to examine general and specific difficulties, as well as advise relatives about the plan of inducing anaesthesia, different approaches, analgesics, and the possibility of further measures such as nasopharyngeal airway insertion postoperatively. Prior to surgery, apart from routine blood investigations, it is essential to do Chest X-ray, 12 lead ECG and rule out cardiac anomalies by doing 2D ECHO.

Anesthesiologist should be prepared with a difficult airway cart in the OT. As is customary, preparedness is a key and child with CLP will require all of the regular paediatric care amenities, as well as those for coping with a possibly difficult airway. In this case scenario, the child had thoraco-lumbar kyphoscoliosis. Kyphoscoliosis can cause breathing difficulties apart from cardio vascular, gastrointestinal and neurological issues (Hedequist & Emans, 2004). So it warrants for thorough preoperative evaluation of cardiovascular as well as respiratory systems. Pulmonary function test is mandatory in these cases. Positioning of the patient for anaesthetic induction is the other major issue in these cases. Due to the hump in the back, proper supine positioning may not be achieved. In such scenarios lateral positioning or placing the hump in a hollow space created by soft cotton rolls can be helpful.

Care should always be taken while giving extended neck position for the surgery. Make sure the head is properly rested and fixed. Using head ring and rolls below the shoulder will help in better positioning. Providing a deep plane of anaesthesia is must. Apart from routine monitoring of ECG, SPO2, ETCO2 and noninvasive BP, intraoperatively strict monitoring should be done on the airway pressures. The possibility of tube kinking/ obstruction is high in CLP surgeries. Detachment of ventilator tubings from ET tube is also common in these surgeries.

Extubation should be done after proper awakening of the patient and after confirming haemostasis. Anesthetist should always examine oropharynx prior extubation to check whether throat packs and blood clots are removed. It is advised to give a lateral positioning to avoid any post nasal dripping of blood and prevent aspiration. Thorough understanding about the patient physiology will help in better anaesthetic management and better outcome.

4. CONCLUSION

Here we successfully managed a case of complete cleft in soft palate with kyphoscoliosis in a 13 year child. Conclusively, a combination of thorough preoperative evaluation with help of multidisciplinary team of experts , vigilant intraoperative monitoring and proper post operative care are essential in such patients.

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Author contributions

Dr.Shiras P: Primary and corresponding author, Data collection and analysis, compilation, case management in the OT
Dr.Sanjot Ninave: Data interpretation, final review of the case report to be published
Dr.Aruna Chandak: Data collection and analysis, compilation, case management in the OT

Dr.Vivek Chakole: Head of the team supervising the case
Dr.Nikhil Bhalerao: Data analysis and interpretation, Conception of or Design of the case report
Dr.Krishnendu S: Data collection, analysis and interpretation

Informed consent
Written & Oral informed consent was obtained from the patient's parents.

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Conflicts of interest
The authors declare that there are no conflicts of interests.

Data and materials availability
All data associated with this study are present in the paper.

REFERENCES AND NOTES

1. Abdelsalam G, Joseph B. Unilateral Cleft Lip and Cleft Palate: A Case Report. Ann Clin Cases 2021; 2(2): 1033.
2. Goswami M, Bhushan U, Jangra B. Management of feeding Problem in a Patient with Cleft Lip/Palate. Int J Clin Pediatr Dent 2016; 9(2):143-5 doi:10.5005/jp-journals-10005-1351
3. Hedequist D, Emans J. Congenital scoliosis. J Am Acad Orthop Surg 2004; 12(4): 266-75. doi: 10.5435/00124635-200407000-00007
4. Hedequist D, Emans J. Congenital scoliosis: a review and update. J Pediatr Orthop 2007; 27(1): 106-16. doi: 10.1097/BPO.0b013e31802b4993
5. Hensinger RN. Congenital scoliosis: etiology and associations. Spine (Phila Pa 1976). 2009; 34(17):1745-50.) doi: 10.1097/BRS.0b013e3181abf69e
6. Jones RG. A short history of anaesthesia for hare-lip and cleft palate repair. Br J Anaesth 1971; 43:796-802. doi: 10.1093/bja/43.8.796
7. Karki SB, Rajbanshi L, Ariyal B, Shrestha K. Anaesthesia for Cleft Lip and Palate Surgery: Study from a Hospital of Eastern Nepal. Birat J Health Sci 2017; 2:127-33 doi: 10.3126/bjhs.v2i1.17288
8. Maharjan SK. Anaesthesia for cleft lip surgery--challenge in rural Nepal. Kathmandu Univ Med J 2004; 2:89-95.
9. Sai Sankar A, Sujatha B, Devi E, Manoj Kumar M. Maiden morsel - feeding in cleft lip and palate infants. J Int Soc Prev Community Dent 2012; 2(2):31-7 doi: 10.4103/2231-0762.109350
10. Sparrow DB, Chapman G, Smith AJ, Mattar MZ, Major JA, O'Reilly VC, Saga Y, Zackai EH, Dormans JP, Alman BA, McGregor L, Kageyama R, Kusumi K, Dunwoodie SL. A mechanism for gene-environment interaction in the etiology of congenital scoliosis. Cell J 2012; 149(2): 295-306. doi: 10.1016/j.cell.2012.02.054
11. Stanier P, Moore GE. Genetics of cleft lip and palate: syndromic genes contribute to the incidence of nonsyndromic clefts. Hum Mol Genet 2004; 13:73-81. doi: 10.1093/hmg/ddh052
12. Takemura H, Yasumoto K, Toi T, Hosoyamada A. Correlation of cleft type with incidence of perioperative respiratory complications in infants with cleft lip and palate. Paediatr Anaesth 2002; 12: 585–8 doi:10.1046/j.1460-9592.2002.00906.x